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CASE REPORT

Dose-dependent relapse of hiatus hernia after administration of intrathecal baclofen treatment—a rare complication

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Abstract
Introduction Intrathecal baclofen treatment (ITB) is widely used in children with cerebral palsy. Although this treatment effectively reduces spasticity, diverse side effects are reported.
Case report We report about a boy with severe asphyxia-induced encephalopathy with bilateral cerebral palsy. After starting the intrathecal baclofen treatment, he episodically showed symptoms of severe gastroesophageal reflux with pale skin color, vomiting, massive drooling, acid regurgitation, and reduced well-being. An open ventral semifundoplication was done some years ago to treat a gastroesophageal reflux. These symptomatic episodes occurred strongly dose-dependent and were not observed during the short test procedure.
Conclusion For the first time, a strong dose-dependent treatment with ITB was documented as a cause for the above episodes and relapsing re-herniations.

Keywords Intrathecal baclofen treatment · Baclofen pump · Cerebral palsy · Gastroesophageal reflux · Semi-fundoplication · Relapse

Introduction

The use of a continuous intrathecal infusion of baclofen by a pump connected to an intrathecal catheter is widely used in the treatment of children with bilateral spastic cerebral palsy (CP) [1]. Parents of children with CP, who have received this treatment, frequently report an increase in quality of life and positive effects regarding the spasticity [2]. Besides the reduction of spasticity, some studies document a weight gain, a reduction of pain, and an improvement in the ease of care after admission of intrathecal baclofen in children with quadriplegia [3, 4]. Even if this therapeutic procedure requires a high initial financial investment, the cost-effectiveness of intrathecal baclofen treatment (ITB) is calculable.

On the other hand, several complications of ITB are reported in numerous case series as well as in some studies, in part device-related such as catheter breakage, malfunctions, or disconnections, and, rarely, pump dysfunctions or complications associated with the therapy procedure such as infections, overdoses, severe withdrawal syndrome in case of malfunction, or hypermobility and hypotonia [5]. In some studies associated with the therapy-induced reduction of the tonus, the progression of scoliosis in patients with bilateral cerebral palsy is discussed [6], although these results are inconsistent [7].

Neurologically impaired children frequently suffer from a gastroesophageal reflux. Several causes are discussed to account for this high incidence of gastroesophageal reflux in children with cerebral palsy such as prolonged supine position, increased intra-abdominal pressure associated with spasticity and/or scoliosis, or hiatus hernia [8].

To our knowledge, and for the first time, we report about a relapse of a hiatus hernia after semifundoplication as a complication of an intrathecal baclofen therapy by a pump.

Case report

Caused by a perinatal hypoxic–ischemic encephalopathy, a 14-year-old boy suffers from severe nonambulatory quadriplegic cerebral palsy (GMFCS level V) associated with symptomatic epilepsy, progressive scoliosis, and spastic

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hip luxation, operated 3 years ago. In addition, the boy suffered from a severe gastroesophageal reflux with regurgitation, vomiting, and failure to thrive beginning at the age of 2. At the age of 6, a hiatus hernia was diagnosed. Despite conservative therapy, a progression of esophagitis was documented. Thereafter, at the age of 7, the boy was operatively treated by an open ventral semifundoplication and percutaneous endoscopic gastrostomy (PEG) tube placement. After this intervention, the gastroesophageal symptoms were significantly reduced.

At 14 years of age, in a test of ITB using a continuous infusion by an external pump, a positive effect on spasticity was documented. During the ITB test, with a dose of 300 μg/24 h, the spasticity was reduced, as was clinically the aim. With respect to this experience and result, caregivers and professionals decided to implant a programmable SynchroMed II pump (Medtronic Inc.) 6 weeks later.

After implantation, the dose was stepwise increased to the aim dose, which successfully decreased the spasticity during the test. Three weeks after reaching the dose of 300 μg/24 h, the boy was acute admitted with pale skin color, vomiting, massive drooling, acid regurgitation, and reduced well-being. The clinical examination and the blood sample, to exclude any metabolic problem or infection, were normal. No malfunction of the pump or dislocation of the catheter was found, and the PEG tube position was adequate. After removing the air from the stomach through the PEG tube and abstinence from food through the tube, the boy recovered slowly over the course of 3 days.

This situation repeated 2 weeks later. The chest X-ray and a single-contrast barium swallow study disclosed a relapse from a large hiatus hernia (Fig. 1). In this situation, the baclofen dose was stepwise reduced to 220 μg/24 h. With this dose, the spasticity was insufficiently reduced. Therefore, two further trials to increase the dose were done, but therapy with an ITB dose of >240 μg/24 h was repeatedly associated with the above-described episodes.

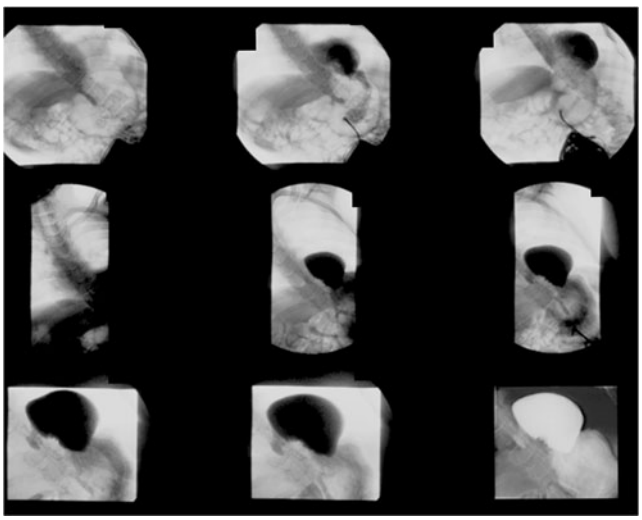

Fig. 1 Single-contrast barium swallow study demonstrating a large hiatus hernia

In an ethical commission discussion, an operative intervention with re-semifundoplication was excluded. Today, the boy is being treated with ITB with 220 μg/24 h—no further clinical episodes of the hiatus hernia, as described above, have been observed over 2 years, and a partial oral feeding is possible, although the spasticity is insufficiently controlled.

Discussion

Even if in an evidence-based review, the data about the efficacy of ITB were judged as insufficient [9]; this treatment is often used, and a lot of clinical reports and studies with small sample sizes report about positive effects in the reduction of spasticity [1]. Consequently, some consensus working groups define a state-of-the art procedure to elect patients who could profit from ITB [10].

Although ITB is a procedure which could have a high grade of multiple benefits for children with CP, there are a number of potential side effects. In up to 31 % of surgical procedures, any complication can occur [5]. In most cases, a device-related complication occurs, rarely with severe consequences. In addition, some nondevice-related adverse effects are described such as increased drooling or relevant hypotonia. Hypotonia induced by ITB is one of the possible causes for the controversially discussed increase of progression of scoliosis after insertion of an intrathecal baclofen pump [6, 7].

Increased intra-abdominal pressure as well as hiatus hernia are possible causes of increased incidence of gastroesophageal reflux in neurologically impaired children. We report about a boy with asphyxia-induced severe bilateral cerebral palsy, who suffered from gastroesophageal reflux disease. After open semifundoplication and PEG tube placement, he showed a stabile course of these gastrointestinal symptoms. However, after implantation of an intrathecal baclofen pump and starting ITB treatment, he developed severe episodes with symptoms of gastroesophageal reflux associated with reduced well-being and vegetative symptoms. A diagnostic work up, including a single-contrast barium study, disclosed a relapse of the hiatus hernia, which was strongly and repeatedly associated with the dose of intrathecal baclofen.

Although recurrent gastroesophageal reflux after fundoplication is reported in up to 46 % of neurologically impaired children [11] and hiatus hernia was identified to be a risk factor for recurrence of gastroesophageal reflux [12], a mechanical breakdown of the hiatus hernia is reported in less than 8 % [13]. In our case, we observed a timely close

connection between a higher dose of intrathecal baclofen and clinical episodes of hiatus herniation. The cause of this relation is unknown. We hypothesize that an increased hypotonia in the trunk causes a sink down of the trunk following increased intra-abdominal pressure. Some additional reports describe that baclofen is effective in treatment of hiccup [14]. The mechanism is unknown. Besides a reduction of the frequency of transient lower esophageal sphincter relaxation, which reduces the frequency of gastrointestinal refluxes as a potential stimulus for hiccup, a direct relaxation of the diaphragm could be discussed. Such a diaphragm relaxation could be a second potential mechanism of the observed hiatus hernia relapse in our case.

In summary, although the external pump trial prior to implantation for intrathecal baclofen in children with CP allows a precise evaluation of effects and safety [15], not all potential complications could be excluded in this test phase. For the first time, with our case, we describe a dose-dependent close connection between ITB and episodes of hiatus herniation.

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